A CASE OF PRIMARY DEGENERATION OF THE PYRAMIDAL TRACTS.*

By William G. Spiller, M.D.,

ASSISTANT CLINICAL PROFESSOR OF NERVOUS DISEASES AND ASSISTANT PROFESSOR OF NEUROPATHOLOGY IN THE UNIVERSITY OF PENNSYLVANIA.

FROM THE WILLIAM PEPPER LABORATORY OF CLINICAL MEDICINE, PHŒBE A. HEARST FOUNDATION.

Spastic spinal paralysis, lateral sclerosis, described separately by Charcot and Erb, has not an uniform pathological basis. Primary degeneration of the pyramidal tracts alone, *i.e.*, a degeneration confined to these tracts, and not resulting from a focal lesion, may occur, but certainly is very rare. The most recent paper on this subject with which I am familiar, is by Ida Democh[2], and she is able to refer only to the cases of Morgan and Dreschfeld[3], and Dejerine and Sottas[4]. She reports a case that is complicated. She speaks of Morgan and Dreschfeld's case as being the least complicated in the literature, and yet some of the cell-bodies of the anterior horns of the spinal cord were diseased in this case The examination of the tissues was made more than twenty years ago when the method of Nissl was not in vogue, and it is highly probable that could this method have been employed the nerve-cell bodies would have appeared more diseased than they did. The case seems to have been one of amyotrophic lateral sclerosis.

In Dejerine and Sottas' case the columns of Goll were not ntact but the nerve cell-bodies of the anterior horns appeared to be normal, although the method of Nissl was not employed.

Ida Democh's case was complicated with neuritis from chronic alcoholic intoxication. The columns of Goll were degenerated in the cervical and upper thoracic regions. The nerve cell-bodies in the anterior horns of the spinal cord were not diminished in number, but apparently the Nissl method was not employed, so that we must remain in doubt whether these cell-bodies were perfectly normal or not. This omission is unfortunate, because in addition to the lateral sclerosis alcoholic neuritis probably existed, and there was therefore a double cause for cellular alteration in the spinal cord.

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One of the most satisfactory cases yet reported is the case given in abstract recently by Strümpell[5]. This patient was 61 years old when first seen by Strümpell, and he was under his observation for almost fifteen years. The case was one of uncomplicated spastic spinal paralysis. The first signs of the disease began in 1866. The rigidity of the lower limbs developed gradually and became intense, and the tendon reflexes were much exaggerated. No fibrillary tremor and no trace of atrophy could be detected. The vesical and rectal functions were not affected. Sensation was normal until near death when some unimportant alteration of sensation was detected. The pyramidal tracts were degenerated, more in the lower thoracic and lumbar regions, and the degeneration did not extend above the pyramids. The columns of Goll were slightly degenerated in the cervical region. The nerve cell-bodies of the anterior horns were perfectly normal. It is presumable that they were examined by the method of Nissl, although the method of examination is not recorded in this abstract. The direct cerebellar tracts may have been slightly degenerated. Strümpell thinks that there can be no doubt that the spastic spinal paralysis from primary degeneration of, and confined to, the pyramidal tracts exists. The upper limbs are not affected in one form of this paralysis, and the disease may be hereditary. Another form is seen in more advanced life, and has a more rapid course, and all the limbs become spastic; finally, there may be slight atrophy of the muscles, so that this form cannot be sharply separated from amyotrophic lateral sclerosis. Two cases of the second type have been studied anatomically by Strümpell.

A third form occurs in childhood and may be hereditary. The pathology of this form has not been determined, except in so far as shown by Bischoff's[6] two cases in brothers, which are not altogether satisfactory. Bischoff found degeneration of the pyramidal tracts as high as the motor decussation, of the columns of Goll, of the direct cerebellar tracts and of the cell-bodies of the anterior horns of the cord and of the motor cortex. These cases cannot be regarded as uncomplicated either from a clinical or pathological viewpoint.

A few cases have been reported in which the primary degeneration of the pyramidal tracts was associated with degeneration of the direct cerebellar tracts, without alteration of the nerve cell-bodies.

My case reported in this paper, has resemblance to Strümpell's[7], published in 1894, in which degeneration of the cell-bodies of the anterior horns existed, although muscular atrophy was not detected during the life of the patient.

A. G., a woman fifty years of age, was admitted to the Philadelphia Hospital, Feb. 26, 1901, complaining of loss of power in the lower limbs, and difficulty of speech. She was at first under the care of Dr. F. Savary Pearce, but later came into my service. The clinical notes, obtained at the time of her admission, were made by Dr. Pearce and the resident physician, Dr. Geisler.

Family history: Her father, mother and one brother were dead from unknown causes.

Personal history: She was the mother of three children, and had had one miscarriage since the birth of her last child. Three years before admission, while coming home from work, she became dizzy, but managed to reach her home without falling. She had not been feeling well for some time. A physician was called and by the time he reached the patient her left upper limb was powerless and she had lost the power of speech. She did not lose consciousness. In a day speech returned, but she has never been able to speak distinctly since the attack. She was able to move her left upper limb within two or three days, and she returned to her work after about three weeks, having recovered except that her speech was still indistinct.

About a year before admission, weakness of the lower limbs was noticed, and this weakness gradually increased in intensity so that she walked very little. Her memory had deteriorated.

On admission she was able to walk with difficulty, and her gait was not distinctly spastic, and her steps were short. She swayed when standing erect, probably from weakness. Her speech was indistinct and bulbar in character.

She was a well-developed muscular woman. Her face was smooth and free from wrinkles below the forehead, but the forehead was wrinkled on both sides. When her mouth was opened it was drawn toward the right side. The naso-labial fold on the left side was not so distinct as on the right. The tongue was protruded straight, but the mouth was not widely opened. Both eyes could be closed. The irides reacted slowly to light and in accommodation. There was apparently no paralysis of the pterygoid, masseter and temporal muscles. There was no disturbance of deglutition.

The chest was well-developed and no atrophy was visible.

The left upper limb was moved freely but was weaker than the right, and the grip of the right hand was stronger than that of the left. There was no atrophy of the hands.

The lower limbs were moved freely, but were weak, and were not atrophied.

The reflexes were as follows:

	RIGHT	LEFT
Biceps-jerk	Much increased.	Increased.
Triceps-jerk	Much increased.	Increased.
Flexors at wrist	Much increased.	Increased.
Extensors	Much increased.	Increased.
Von-Bechterew's	Absent.	Absent.
Epigastric	Absent.	Absent.
Knee-jerk	Much increased.	Increased.
Quadriceps-jerk	Much increased.	Increased.
Achilles-jerk	Present.	Present.
Plantar reflex	Increased	Increased.
Babinski's	Extension of great toe.	Slight extension of great toe.
Ankle-clonus	Absent.	Absent.
Patellar clonus	Absent.	Absent.

There were no disturbances of sensation, and stereognostic perception was normal.

She had been constipated and had noticed an increasing difficulty in retaining the urine for five months, but had not had absolute incontinence.

She had had dull headache over the whole of the head for one year, and occasionally had had pain in the left side of the face and in the back, but she had not had pains in the limbs.

On March 3, 1901, Dr. Charles A. Oliver examined her eyes and reported: "Left pupil is the larger. The irides are extremely sluggish to light, particularly the left one; quite prompt to accommodation and convergent efforts. Paresis of left external rectus muscle. Eyegrounds are healthy."

An urinary examination made March 1, 1901, gave the following results: "Lemon color, sp. gr. 1011, reaction acid, sediment white; microscopical examination: pus and epithelial cells; chemical analysis: albumin, small amount; no sugar."

Another examination made July 8, 1901, showed granular casts and epithelial cells, and a large amount of albumin.

On July 3, 1901, the following notes were made by me: The patient is in a stuporous condition, although she replies to questions and moves her limbs on command. She permits the flies to collect on her face, and even on the inner side of the lips or within the mouth, without attempting to brush them away. She can give her name, but is unable to give her age, or to say how long she has been in the hospital. Her speech is thick. She has incontinence of urine and feces. She is paretic in the lower limbs, though she can move all the muscles of these limbs. The lower limbs are not atrophied and are flaccid.

In testing sensation no dependence can be placed on her statements, but she recognizes the point of a pin and withdraws either lower limb when it is stuck with a pin.

The knee-jerk on the right side is diminished, and is less intense than that on the left, which is also impaired. The Achilles-jerk on the right side is present, but feeble; on the left side it cannot be distinctly obtained. The plantar reflex is exceedingly active on each side, and the Babinski reflex is pronounced on each side. [A few days later the Babinski reflex was absent on each side, and no movement of the toes was obtained by irritation of the sole of the foot. The loss of this reflex was probably a result of the increasing stupor.]

She moves the upper limbs freely, and these limbs are not distinctly paretic. There is no distinct atrophy of the upper limbs. When the upper limbs are stuck with a pin she shows signs of discomfort.

The pupils are unequal, the left being larger than the right. The movements of the eyeballs seem to be free in all directions. The irides contract feebly to light; there is very little contraction in convergence. She closes the eyelids firmly. Both upper lids droop so that the palpebral fissures are narrow.

The heart sounds are very loud, and the first sound at the apex beat is suggestive of a murmur, the murmur however is not transmitted into the axilla. The second pulmonic sound is somewhat accentuated.

The patient became progressively weaker, pulmonary and cardiac disturbance and fever developed, and she died July 10, 1901.

The pathological diagnosis made by Dr. W. F. Hendrickson, July 11, 1901, was: "Acute lobar pneumonia, chronic interstitial nephritis, hypertrophy and dilatation of heart, general arteriosclerosis, localized chronic pericarditis, chronic endocarditis, parenchymatous degeneration of myocardium, edema and congestion of lungs, early acute splenic tumor, parenchymatous degeneration of liver, chronic gastritis."

The brain and spinal cord were examined by me. Microscopical sections were made from the left paracentral lobule, the right cerebral peduncle, pons and lower parts of the central nervous system.

A slight recent hemorrhage was found within the cortex of the left paracentral lobule. The cell-bodies of this lobule could not be well studied by the thionin stain. No meningitis was found over this lobule, but the blood vessels of the pia at this part were slightly thickened.

The foot of the right cerebral peduncle was not degenerated either when studied by the Weigert hematoxylin or the Marchi stain. The right oculomotor nucleus contained many nerve cell-bodies and nerve fibers. A group of thickened blood vessels, surrounded by round-cell infiltration and the remains of an old hemorrhage, was found within the right cerebral peduncle near the oculomotor nuclei. No meningitis was found at the foot of the peduncle, and the blood vessels of the pia at this part were not much thickened.

The motor tracts within the pons were very slightly degenerated. The nucleus of each sixth nerve seemed to be normal. Very slight degeneration by the Marchi stain was found in the right pryamid, and none was found by this stain in the left pyramid. The cell-bodies of the nucleus of each twelfth nerve were shown to be deeply pigmented by the Marchi stain. The cell-bodies of these nuclei were numerous, but possibly not so numerous as in normal sections, and so far as could be determined by imperfect thionin staining they were not much diseased. The intramedullary portions of the twelfth nerves were normal. No meningitis was found over the medulla oblongata, and the vessels of the pia were not notably thickened. The anterior pyramids were slightly degenerated as shown by the Weigert hematoxylin stain, and the left pyramid was a little more degenerated than the right.

The degeneration of the pyramidal tracts in the spinal cord, as shown by the Marchi method, was very slight, but when the Weigert hematoxylin stain was employed the crossed pyramidal tracts in the lower cervical and lumbar regions were found distinctly but by no means completely degenerated, and the degeneration was therefore of long standing. It was equal on the two sides of the cord. The direct cerebellar tracts were normal The posterior columns were apparently intact. The direct pyramidal tract on the left side in the cervical region was slightly degenerated. The cell-bodies of the anterior horns in the lower cervical region were not so numerous as in normal sections, and some of these cell-bodies were diseased; *i.e.*, some were shrunken and had lost their chromophilic elements. The anterior roots of the lower cervical region appeared to be normal; except that some of the axones may have been slightly swollen.

In the lumbar region the cell-bodies of the anterior horns were not so numerous as in normal sections, and some of these cell-bodies were diseased in the same way as those of the cervical region. The anterior roots seemed to be normal.

Summary.—A woman fifty years of age developed suddenly weakness in her left upper limb with loss of speech. The power of speech was regained after a day or two, but never again was normal. The weakness almost entirely disappeared from the left upper limb after about three weeks. Two years after this attack she noticed that she was weak in her lower limbs, and the weakness gradually increased so that walking became difficult. The reflexes in all the extremities were exaggerated, and the Babinski reflex was obtained. The gait was not decidedly spastic. The right upper limb was not distinctly paretic, but the left was a little weak. No objective sensory disturbances were found, and no pains were felt in the limbs. Muscular atrophy was not observed, although the palsy of the lower limbs had existed more than a year. The reaction of the irides was sluggish. The left side of the face was slightly paretic, but the right side also probably was not normal.

Degeneration of the pyramidal tracts was found extending as high as the pons, but not above this. This degeneration was less intense in the anterior pyramids than in the spinal cord, and was equal on the two sides of the cord. The other tracts in the cord were normal. No meningitis was present.

Some of the small vessels of the cerebral pia were thickened, which in a person fifty years of age was not remarkable. The cell-bodies of the anterior horns of the spinal cord were in part diseased, and the nuclei of the hypoglossal nerves were probably not absolutely normal.

The attack of paralysis of the left upper limb probably with implication of the face, and with speech disturbances which developed two years before the other symptoms, cannot be satisfactorily explained by the findings. Amyotrophic lateral sclerosis does not usually begin in an apoplectiform manner. The degeneration of the pyramidal tracts extended equally high on the two sides, so that it is not probable that a focal lesion in the cerebrum was the cause of the degeneration in either tract; and no focal lesion could be found. A small hemorrhage in the brain may have occurred and caused the paralysis, but if so it did not lead to secondary degeneration. It seems not at all impossible that the sudden weakness of the left upper limb and of the left side of the face was the first sign of the amyotrophic lateral sclerosis, and that the weakness in the lower limbs developed so gradually that it caused little annoyance to the patient until a year or two later.

A case of amyotrophic lateral sclerosis with necropsy reported by Schlesinger[8] has a resemblance to my case. A man, seventy-two years old, received a severe mental shock and his speech became at once affected. Soon after this a temporary right-sided hemiparesis developed. The mouth was opened with difficulty and deglutition was affected. To the symptoms of bulbar palsy were added progressive spastic paresis, without distinct atrophy of the extremities, and with exaggeration of the tendon reflexes. An acute commencement or rapid progression of bulbar palsy, according to Schlesinger, should always suggest the possibility of amyotrophic lateral sclerosis.

The paralysis in my case beginning in the lower limbs and not implicating the upper limbs, except the persisting slight paresis in the left upper limb, dating from the apoplectic attack, is unusual in amyotrophic lateral sclerosis, but has been described as one of the signs of the spastic paralysis of Erb and Charcot from primary degeneration of the pyramidal tracts. The gait was not very spastic but it was weak and the tendon reflexes were exaggerated.

The absence of distinct muscular atrophy was one of the most interesting features of this case. The disease had existed more than a year, and therefore time for atrophy to develop had been given, and yet even in the lower limbs distinct atrophy was not observed. This is especially noteworthy, inasmuch as some of the cell-bodies of the anterior horns of the spinal cord were diseased. Had death been delayed atrophy might have been observed. It seems probable that the cellular changes developed later than the degeneration of the pyramidal tracts.

In the weakness of the lower limbs, the exaggerated reflexes without muscular atrophy, the gradual development of the paralysis—gradual because at the time of admission to the hospital the patient was still able to walk—the case presented the clinical picture of primary lateral sclerosis, and yet the microscopical examination showed that the cell-bodies of the anterior horns were implicated.

The slow reaction of the irides is not common in amyotrophic lateral sclerosis, but Schlesinger[8] has observed the Argyll-Robertson phenomenon in this disease. The slow reaction in my case may have been caused by anteriosclerosis.

Amyotrophic lateral sclerosis is usually regarded as a rare disease, and yet within the last three or four years I have studied the pathological material from five cases of this affection, but only two of these cases have been reported. I have seen quite a large number of clinical cases within this period, so that I am inclined to think that amyotrophic lateral sclerosis is not so rare as many physicians believe. In association with Dr. Dercum[9], and later in quite a long paper by myself[10], I have discussed the symptomatology and pathology of this disease, and it is hardly worth while to repeat what has been said. In only one of the five cases was I able to trace degeneration above the pons, and in that case the degeneration of the motor cortex was so intense that I believed that by the method of Marchi I might be able to define the extent of the cortical motor area, inasmuch as amyotrophic lateral sclerosis is essentially a disease of the motor system; an attempt which had never been made. This I was able to do, and I obtained a motor area corresponding quite closely to that described by von Monakow and others. The ascending frontal convolution was more degenerated than the ascending parietal, and this finding is especially interesting in connection with the recent results obtained by Schaffer[11] in his study of brains from cases of paretic dementia, and by Sherrington and Grünbaum[12] in their experiments on the brains of monkeys. These studies seem to show that the motor functions are represented in the ascending frontal convolution much more than in the ascending parietal convolution.

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[2] Ida Democh, Archiv. f. Psychiatrie, Vol. 33, No. 1, p. 188.

[3] Morgan and Dreschfeld, British Med. Journal, Jan. 29, 1881.

[4] Dejerine and Sottas, Arch. de physiol. norm. et path., 1896, p. 630.

[5] Strümpell, Neurologisches Centralblatt, July 1, 1901, No. 13, p. 628.

[6] Bischoff, Wiener klin. Woch., No. 51, Dec. 19, 1901, p. 1281.

[7] Strümpell, Deutsche Zeitschrift f. Nervenheilkunde, Vol. v, p. 225.

[8] Schlesinger, Arbeiten aus dem Neurologischen Institute an der Wiener Universität. Vol. vii, 1900.

[9] Dercum and Spiller, JOURNAL OF NERVOUS AND MENTAL DISEASE, Feb., 1899.

[10] Spiller, "Contributions from the William Pepper Laboratory of Clinical Medicine," 1900, and author's abstract in JOURNAL OF NERVOUS AND MENTAL DISEASE, March, 1900, p. 165.

[11] Schaffer, Neurologisches Centralblatt, No. 2, Jan. 16, 1902, p. 54.

[12] Sherrington and Grünbaum, British Medical Journal, Vol. 2, 1901, p. 1091.